



Double-orifice mitral valve in an asymptomatic young adult woman with aortic coarctation and bicuspid aortic valve

Doble orificio de válvula mitral en mujer asintomática joven con coartación aórtica y válvula aórtica bicúspide

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Keywords:

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Palabras clave:

Doble orificio mitral, malformaciones congénitas, técnicas de imagen multimodal.

ABSTRACT

Congenital heart disease in Mexico ranks second among congenital malformations in newborns, with an incidence of 0.8-1.4%. Double orifice mitral valve (DOMV) is rare congenital heart disease, with a reported incidence of 0.05%. This malformation consists of the anatomical presentation of two mitral orifices, commonly associated with other congenital malformations such as septal defects, complete or partial malformations of the atrioventricular canal, aortic coarctation, tetralogy of Fallot, atrial or interventricular communication, Ebstein's anomaly and patent ductus arteriosus. The clinical characteristics are variable and can even go unnoticed and be diagnostic until adulthood. We present the case of an asymptomatic young woman with a diagnosis of arterial hypertension. During a routine control was diagnosed with multiple congenital heart diseases whose relationship is little described in the literature, this being the third case reported. At present, multimodal imaging techniques allow a greater characterization of lesions of both the valve and the valve apparatus, intending to carry out a comprehensive diagnostic and therapeutic approach to offer the greatest benefit to the patient.

RESUMEN

Las cardiopatías congénitas en México ocupan el segundo lugar de malformaciones congénitas en los recién nacidos, con una incidencia de 0.8-1.4%. El doble orificio de la válvula mitral (DOVM) es una cardiopatía congénita rara, con una incidencia reportada de 0.05%. Esta malformación consiste en la presentación anatómica de dos orificios mitrales comúnmente asociada a otras malformaciones congénitas como defectos septales, malformaciones completas o parciales del canal atrioventricular, coartación aórtica, tetralogía de Fallot, comunicación interauricular o interventricular, anomalía de Ebstein y persistencia del conducto arterioso. Las características clínicas son variables e incluso pueden pasar desapercibidas y ser diagnosticadas hasta la edad adulta. Se presenta el caso de una mujer joven el cual se encuentra asintomática con diagnóstico de hipertensión arterial, que durante una revisión de rutina fue diagnosticada con múltiples cardiopatías congénitas, de las cuales, su relación está poco descrita en la literatura, siendo este el tercer caso reportado. En la actualidad las técnicas de imagen multimodal permiten la mayor caracterización de las lesiones tanto de la valva como del aparato valvular, con la intención de realizar un abordaje diagnóstico y terapéutico integral para ofrecer el mayor beneficio al paciente.

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INTRODUCTION

The double-orifice mitral valve (DOMV) is a rare congenital disease first described in 1876 by Greenfield^{1,2}. DOMV has been associated with atrioventricular septal defects, partial or complete atrioventricular canal malformations, aortic coarctation, tetralogy of Fallot, atrial or ventricular septal defect, Ebstein's anomaly, and patent ductus arteriosus. We present this case to add relevant evidence to the use of multiple imaging modalities for congenital heart diseases, particularly for complex valvular heart disease.

CASE PRESENTATION

A 29-year-old woman with a 5-year history of hypertension who presented without symptoms since diagnosis. During the control and physical examination, a systolic murmur was found in the left parasternal border as well as a systolic-diastolic murmur in the interscapular area with decreased femoral pulses; CT angiography revealed a coarctation of the thoracic aorta near the left subclavian artery (Figure 1).

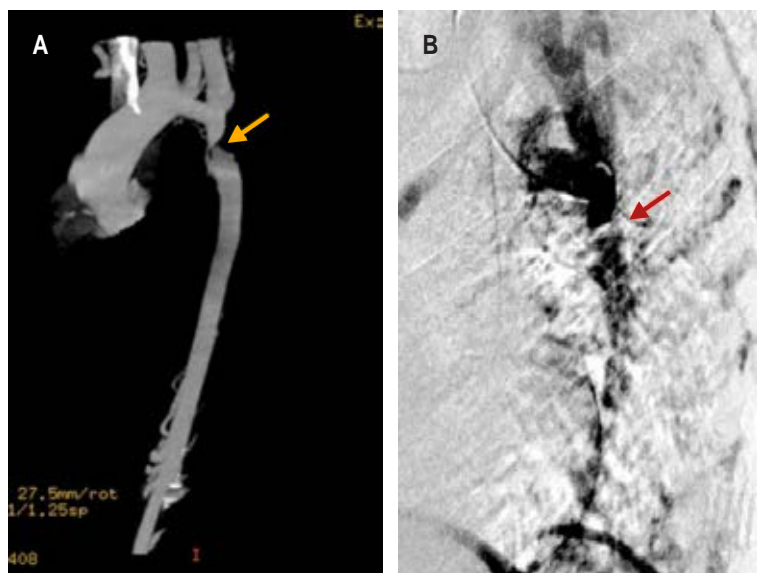


Figure 1: Reconstructed CT aortogram showing localized narrowing of the aorta. **A)** Reconstruction of the coronal angio-tomography, the projection shows the coarctation site that is 21 mm from the left subclavian artery. **B)** Digital subtraction angiography showing the thoracic aorta with coarctation.



Figure 2: 2D transesophageal echocardiography at the mild esophageal projection at 57° showing the bicuspid aortic valve.

Echocardiography was performed using the ACUSON SC2000™ ultrasound system (Siemens, Munich, Germany), which showed a bicuspid aortic valve (Figure 2) with moderate stenosis (maximum velocity of 3.4 m/s, maximum gradient of 31 mmHg) and a double-hole mitral valve with thickened valves, insufficient chordae tendineae and two papillary muscles. The posteromedial muscle was hypoplastic and two cords were inserted into the anterolateral muscle, with a mean gradient of 4 mmHg, a mitral valve area of 1.3 cm², and a severely insufficient eccentric jet through the posterolateral wall of the atrium left with the Coanda effect. The mitral valve orifices were a small orifice in the middle and a larger lateral orifice, which according to the classification of Baño-Rodrigo, can be classified as type 5 or type 2 and is not associated with defects of the atrioventricular canal³⁻⁵ (Figure 3). A CMR was performed to confirm the bicuspid aortic valve and double-hole mitral valve (Figures 4 and 5). The patient underwent surgical mitral valve replacement and was discharged without complications after five days. One month after surgery, the patient was asymptomatic in NYHA class I.

DISCUSSION

Aortic coarctation has been reported to be associated with bicuspid aortic valves in 30 to 80% of cases; in the present case, the patient also had a congenital malformation

of the mitral valve, which is rare and occurs in only 0.05% of all congenital heart diseases. DOMV has been widely associated with septal defects and with a complete or partial atrioventricular canal in 0.05% of cases.⁶ In this particular case, in addition to the aortic coarctation and the bicuspid valve, the patient was asymptomatic; This association

has been rarely described in the literature, and as far as we know, this is the third case reported to date.

DOMVs can be anatomically classified into central, valvular, and commissural categories according to their structures or conditions; DOMV can also be classified by two-dimensional echocardiography as follows: 1) complete bridge (15% of cases), both orifice openings are circular and visible from the edge of the valve, with normal subvalvular system and papillary muscles; 2) incomplete bridge, missing valve connection and can only be visualized on the edge of the valve; and 3) hole type (most common), typically a minimally sized accessory hole in the posteromedial or anterolateral commissure.^{7,8} In this particular patient, the VMOV was diagnosed as type 2 according to the Baños-Rodrigo classification, since the accessory orifice (the smallest) is located in the posteromedial commissure.⁵

Accessory holes are made by different mechanisms that result in DOMV:⁵

1. Ring of strings
2. Subdivision of the muscular crest
3. Fused papillary muscle
4. Crossing of strings
5. Central fibrous subdivision

The bicuspid aortic valve has not been related to DOMV, and although its coexistence has been described in up to 50% of cases, there is no relationship in adults; therefore, this case presents this rare anomaly and associated congenital malformations.

This case exemplifies the importance of the different cardiac imaging techniques since the patient was referred for probable aortic coarctation. The CT aortogram was the initial imaging study performed. Then a transthoracic echocardiogram was performed; because the bicuspid aortic valve and DOMV were found, MR was necessary and useful to determine the timing and definitive surgical treatment.

CONCLUSIONS

DOMV is a rare congenital malformation that can be associated with structural cardiac

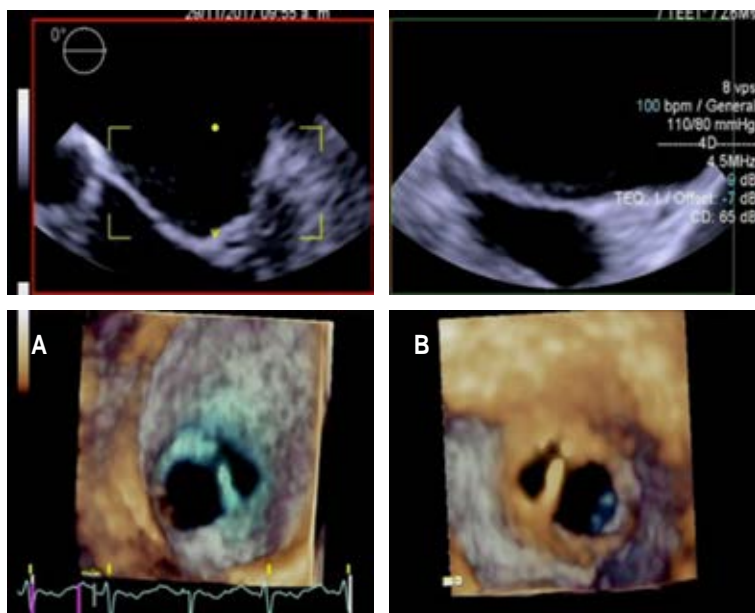


Figure 3: Real-time 3D transesophageal echocardiography of **A** atrial face and **B** ventricular face. It is an eccentric type of double port mitral valve with a larger main port and a smaller accessory port located at the anterolateral commissure.

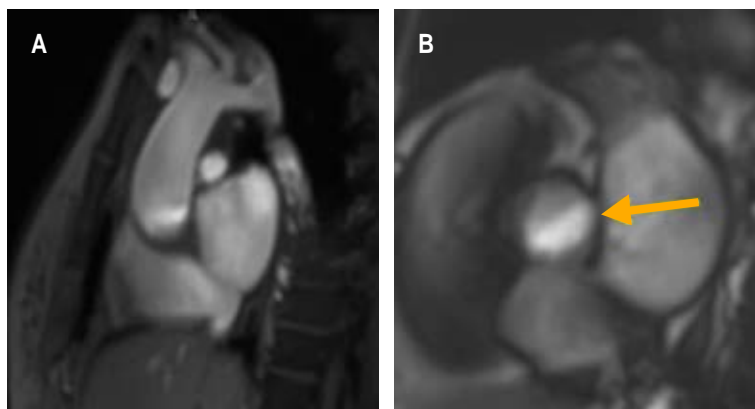


Figure 4: **A)** A rendered image of the 3D contrast magnetic resonance angiogram showing the thoracic aorta. **B)** Cine CMR image of the bicuspid aortic valve in systole (yellow arrow).

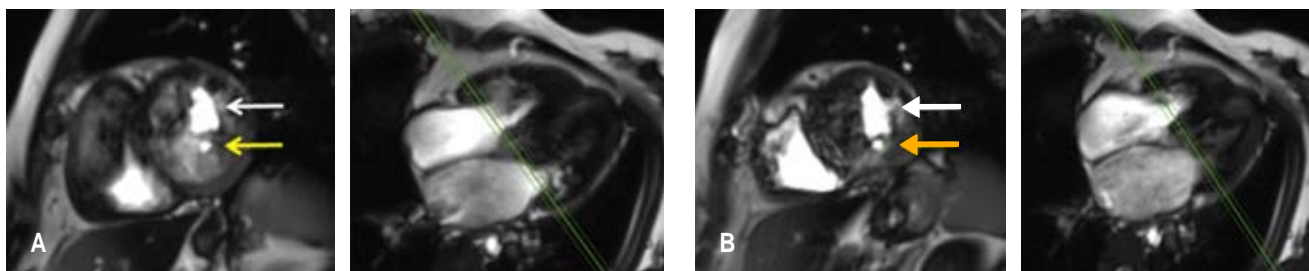


Figure 5: Cardiac magnetic resonance image (Siemens Verio 3T). **A)** Image acquired with the SSFP sequence showing the mitral valve in diastole and the double orifice of the mitral valve (white and yellow arrows). Along the short axis, where the tips are in diastole, the image shows the double mitral orifice, where the posterior orifice is the most minor (yellow arrow) with an area of 0.17 cm^2 , and a smaller accessory orifice it is located anterolaterally commissure. **B)** Images of the SSFP sequence along the short axis at the apical level of the double mitral foramen (white and yellow arrows). Parallel green dashed lines indicate the cut-off position to image valve tips to assess the valve area.

abnormalities. We have presented strong evidence for the use of multiple imaging modalities to fully assess congenital heart disease and the added value of 3D transesophageal echocardiography.

REFERENCES

1. Greenfield WS. Double mitral valve. *Trans Pathol Soc London*. 1876; 27: 128-129.
2. Kim SJ, Shin ES, Lee SG. Congenital double-orifice mitral valve with mitral regurgitation due to flail leaflet in an elderly patient. *Korean J Intern Med*. 2005; 20 (3): 251-254.
3. Izaguirre CE, Valiente MJ, Desdín HY, Valiente TA, Turro FMA. Válvula mitral de doble orificio (VMDO) con anomalía cardíaca asociada a aorta bicúspide y coartación de la aorta en pacientes pediátricos. A propósito de dos casos. *Revista Científica Hallazgos*. 2017; 2 (2): 1-6.
4. Sharma S, Loya YS, Daxini BV. Coarctation of aorta with unusual association of diverticulum of the left ventricle and double orifice mitral valve. *Int J Cardiol*. 1991; 30 (1): 113-115.
5. Baño-Rodrigo A, Van Praagh S, Trowitzsch E, Van Praagh R. Double-orifice mitral valve: a study of 27 postmortem cases with developmental, diagnostic and surgical considerations. *Am J Cardiol*. 1988; 61 (1): 152-160.
6. Han J, He Y, Li Z, Zhang Y, Chen J, Wang L et al. Isolated double-orifice mitral valve anomaly on 3-dimensional transesophageal echocardiography. *J Ultrasound Med*. 2009; 28 (11): 1589-1592.
7. Banerjee A, Kohl T, Silverman NH. Echocardiographic evaluation of congenital mitral valve anomalies in children. *Am J Cardiol*. 1995; 76 (17): 1284-1291.
8. Laccetta G, Napoli AM. Incidental finding of an isolated double-orifice mitral valve in an asymptomatic man. *J Cardiovasc Echogr*. 2016; 26 (3): 100-103. doi: 10.4103/2211-4122.187963.

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